Results: We identified 38 patients, with the diagnosis of AAV made 11 months to 19 years previously. 16 (42%) had been diagnosed within the last five years. Age range was 19-87 years old with roughly half (21; 54%) being male. Maintenance Rituximab was started between January 2010 and September 2018. Most patients received a fixed 6 monthly protocol. Seven patients (18%) died within the observation period. More details in table 1 below.

Table 1. Results

Total number (n) patients = 38	n (%)
Serology	32 (84.2%)
ANCA positive	21 (55.3%)
PR3 positive	9 (23.7%)
MPO positive	
Previous cyclophosphamide (2/38 unknown)	20/36 (55.6%)
Indication for starting Rituximab	9 (23.7%)
Failure of cyclophosphamide	20 (52.6%)
Flare/worsening disease despite disease-modifying therapy	3 (7.9%)
Intolerant of other disease-modifying therapies	3 (7.9%)
Young patient of child-bearing potential	2 (5.3%)
Patient preference	1 (2.6%)
Not documented	
Concurrent medication	12 (31.6%)
Azathioprine	11 (26.3%)
Mycophenolate mofetil	3 (7.9%)
Methotrexate	1 (2.6%)
Hydroxychloroquine	1 (2.6%)
Tacrolimus†	12 (31.6%)
Prednisolone monotherapy	1 (2.6%)
Nil	
Cotrimoxazole	20 (52.6%)
Prescribed	2/20 (10%)
Intolerant	
Infections	18 (47.4%)
Documented in clinic letters	13 (34.2%)
Any infection	10 (26.3%)
Respiratory tract infection	
Requiring admission	
Immunoglobulins post-Rituximab	4 (10.5%)
Hypogammaglobulinaemia	1/4 (25%)
Of which treated with IV immunoglobulins	3 (7.9%)
Low IgG and IgM	17 (44.7%)
Isolated low IgM	3 (7.9%)
Not tested	
Clinical response to Rituximab	31 (81.6%)
Full	2 (5.3%)
Partial; requiring addition of further disease-modifying therapy	2 (5.3%)
Mild disease progression	3 (7.9%)
Flare within first 2 years of treatment‡	
First 2 years of Rituximab treatment	11 (28.9%)
Ongoing	24 (63.2%)
Completed	3 (7.9%)
Not completed	1 (2.6%)
Discontinued due to infections	2 (5.3%)
Patient died	
Subsequent treatment ^Y (n=24)	6/24 (25.0%)
Rituximab continued as primary therapy	6/24 (25.0%)
Regular Rituximab restarted following flare	1/24 (4.2%)
Mycophenolate mofetil	4/24 (16.7%)
Azathioprine	2/24 (8.3%) 5/38 (13.2%)
Prednisolone monotherapy Patient died	

† Renal transplant patient ‡ In 2/3 (67%) patients associated with poor compliance/missed doses

^Y In patients who completed 2 years maintenance Rituximab

Conclusion: The effectiveness of Rituximab as a maintenance therapy in AAV is borne out by this real world data, with 31/38 patients demonstrating full clinical response. Longer term treatment and low grade disease continue to be issues as evidence for optimal management strategies is lacking. Infections, including severe infections necessitating admission, were common. PR3 positive patients are over-represented, in keeping with the known higher rate of relapse.

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I confirm that the abstract fulfils the requirements for a late breaking submission and that the research results were not available at the time of the original deadline (31 January 2019)

Yes

Disclosure of Interests: Megan Rutter: None declared, Shirish Dubey Grant/research support from: Roche provided a grant 5 years ago, Speakers bureau: Yes, BMS and Internis, Andrew Short: None declared
DOI: 10.1136/annrheumdis-2019-eular.2364

AB0632B TB OR NOT TB, THAT IS THE VASCULITIC QUESTION

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Background: It is important to remember that there are many disorders that can mimic the clinical, radiographic and histological features of vasculitis. Tuberculosis (TB) is often referred to as the great imitator and therefore, needs to be considered early in the differential diagnosis of vasculitis.

Objectives: To emphasize the importance of being aware of noninflammatory mimics of vasculitis to be able to avoid unnecessary and potentially harmful immunosuppression.

Methods: We report a challenging case of a patient presenting with an inflammatory subglottic lesion.

Results: A 81-year-old lady initially presented to otorhinolaryngology (ENT) with hoarseness and restriction of breathing. Nasendoscopy revealed a subglottic lesion with oedema and inflammation. Biopsy showed florid necrotising granulomatous inflammation. Upon questioning, patient revealed one year history of epistaxis and nasal crusting requiring regular nasal douching. She also reported lack of energy, loss of appetite, but denied any rash, weight loss or night sweats. She suffered with a severely dry mouth that was causing difficulty swallowing. She also complained of an occasional cough with small amount of phlegm which she attribute to the difficulty with swallowing. Rheumatology team felt that this could be a possible localised Granulomatosis with polyangiitis and requested ANCA and a vasculitis screen.

Interestingly, the patient's chest x-ray showed apical scarring, consistent with healed mycobacterial disease. A CT neck/thorax and abdomen was done which showed diffuse mucosal thickening of the subglottis and reticulonodular changes in the lungs. She was also noted to have deranged liver function test around this time. Gastroenterology advised that there was no clear cause for the transaminitis and an autoimmune screen was requested. Given the chest findings on CT, a sputum sample was sent for microscopy and culture. The patient had been started on dexamethasone on admission by ENT and reported substantial improvement in the hoarseness of voice and shortness of breath. ANCA then came back negative, as did the connective tissue screen. While the rheumatology team was considering how to further manage the possible vasculitis, the sputum smear came back positive for acid fast bacilli and the diagnosis was clinched: our patient was suffering with the extremely rare laryngeal tuberculosis, along with pulmonary TB. She was started on anti-tuberculous medication that have been introduced incrementally given the deranged liver function tests. The patient was doing well on her most recent follow up in the respiratory department.

Conclusion: Greater awareness of vasculitis mimics like TB and a high index of suspicion are needed when assessing a patient presenting with the protean manifestations of suspected vasculitis. It is therefore prudent to include TB in the differentials, whenever consider vasculitis.

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Abstract AB0632B Figure 1: View of the larynx on endoscopy showing the suspicious nodules of granulomatous inflammation

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.8260

Scleroderma, myositis and related syndromes

AB0633

FECAL CALPROTECTIN IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a connective tissue disease with heterogeneous symptoms and a non-predictable process. The most involved organ after the skin is the gastrointestinal tract (GI tract) and any part of it can be affected. The calprotectin level is increased in plasma, urine, sputum and feces when an inflammatory process is occurring with the presence of neutrophils which is measurable.

Objectives: So we decided to examine fecal calprotectin as a simple method to diagnose GI disorders and disease activity in SSc.

Methods: Totally 46 patients with SSc were invited by telephone, who have been referred to sayyad shirazi Hospital and the rheumatologists' offices in Gorgan and their information has been registered there. Seven patients did not enter our study because of having the items in exclusion criteria like Diabetes, a history of GI operation and having other connective tissue diseases at the same time. For the other 39 patients who was remained, a questionnaire about GI symptoms (UCLA SCTS 2.0) was filled in and then their skin was examined and the skin score was determined (the score more than 14 was considered as positive). Blood sample was taken from all patients to do tests as ESR, CRP, Magnesium, Albumin, Ferritin, Folic Acid, Vitamin B12 and Zinc and use them as a scale for malnutrition. Then the stool samples were collected to measure the calprotectin level. At last to determine GI disorders in a non-aggressive way, Barium Swallow (Esophagus malfunction) and CT Enterography (intestinal disorders) was done. The patients in our study according to the median of age which was 42, was divided in to two groups: young and middle aged. Due to the fact that FC level wasn't less than 50 µg/gr in any of our cases, the patients was divided in to two groups: the one with FC level less than 200 µg/gr and the one with FC level as 200 µg/gr or more than 200 µg/gr and the latter was considered as positive.

Results: As we didn't have any male patients, the correlation between FC level and sex was not verifiable. Increased levels of FC didn't have any correlation with age (P Value=0.79), disease type (P Value=0.59), small intestine wall thickening (P Value=0.10), esophageal dilatation (P Value=0.18), skin score (P Value=0.25), final GI score (P Value=0.30) and none of serum tests. One of the most important reasons, was low sample size. In our study just 5 cases had evidence of wall thickening in CT Enterography and all that 5 cases had increased levels of calprotectin so we see that if we have a larger sample size we may ensure the

connection between these two variables. Also as most of our patients take vitamin and mineral supplements, we couldn't find any signs of micronutrient deficiencies and any correlation with FC level as well.

Conclusion: Although there was no statistical correlation between FC level and variables, the results are in favor of FC's specificity for assessing intestinal wall thickening.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.7663

AB0634

FINDINGS OF EXERCISE ULTRASOUND CARDIOGRAPHY IN PRE-PAH PATIENTS WITH SYSTEMIC SCLEROSIS ARE CORRELATED WITH EXTENT OF CAPILLARY LOSS DETECTED BY NAILFOLD VIDEOCAPILLAROSCOPY

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Background: Systemic sclerosis (SSc) is characterized by excessive fibrosis, injuries in vasculature, and production of autoantibodies (1). Patients with SSc are complicated with various organ involvements, which can give a great impact on their prognosis (2). Of these, pulmonary arterial hypertension (PAH) is one of leading causes of death and the determinant of prognosis in SSc (3). And early diagnosis is an important key to improve their prognosis. The nailfold videocapillaroscopy (NVC) is a non-invasive modality which can evaluate the status of microvasculature of patients with SSc. However, the significance of NVC in patients with "pre-clinical" PAH was unclear.

Objectives: To examine the significance of detection of microvascular injury by NVC in patients with pre-clinical PAH patients with SSc.

Methods: Patients with SSc who newly visited the outpatient clinic at Fujita Health University Hospital between August 2015 to March 2016 were consecutively involved. All the patients fulfilled the 1980 preliminary classification criteria for SSc (4). The diagnosis of PAH was based on the definition in 2015 ESC/ERS guideline. The NVC (Medcap[®]3.0, DS MEDICA) was used to detect microvascular change and average capillary density and the number of capillaries/view were calculated. Also, existence of giant capillaries, hemorrhage and patterns of capillary changes were also documented. Transthoracic ultrasound cardiography (UCG) was performed using iE33 with 5MHz sector probe S5-1 (Philips). Exercise loading was done with an ergometer (Lode). Ejection fraction, E/e', velocity of tricuspid regurgitation jet (TR jet), tricuspid annular plane systolic excursion (TAPSE) were examined in a supine position. Tricuspid regurgitation pressure gradient (TRPG) was calculated using velocity of TR jet based on Bernoulli principle. Exercise loading was started at 25 Watt, and then increased 25 Watt every 3 minutes and TRPG at maximum exercise was also calculated. Parameters of NVC were compared with those of UCG. Patients with "pre-clinical" PAH were defined as patients 1) asymptomatic, 2) who did not meet the definition of PH by right heart catheterization (RHC), 3) whose probability of PH is lower than "intermediate" by UCG, 4) who did not see any UCG signs for PH based on the guideline and 5) who did not have severe interstitial lung disease, especially% FVC > 70% and/or area of ILD based on CT scan was <20% of the entire lung field.

Results: Nineteen patients with SSc were newly visited. Male to female ratio was 4:15, mean age 63 ± 13 years, and mean disease duration was 138 ± 109 months. Ratio of limited skin type was 79%. 37% were with ILD and 11% were with PAH confirmed by RHC. There is no correlation between TRPG at rest and the density of capillaries. However, there is a significant, negative correlation between TRPG at exercise and the capillary density ($r = 0.57$, $P < 0.02$). Furthermore, the same result was observed even with seventeen "pre-PAH" cases ($r = 0.67$, $P < 0.04$).